Vol. 72, No. 9 Established 1884 September 1955

ARCHIVES OF PEDIATRICS

A MONTHLY DEVOTED TO THE

DISEASES OF INFANTS AND CHILDREN

JOHN FITCH LANDON, M.D., Editor

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E. B. TREAT & CO., Inc., *Publishers*, 45 East 17th Street, NEW YORK, 3
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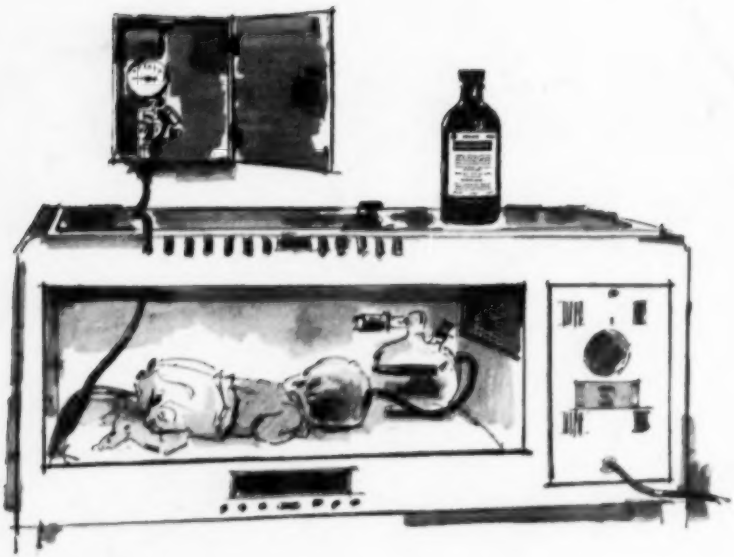

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Editorial Communications address to JOHN F. LANDON, M.D., 120 East 75th Street, New York 21

Business Communications address to E. B. TREAT & Co., INC., 45 East 17th Street, New York 3

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*Farley, W. J.: Oxytetracycline in Pediatrics, Internat. Rec. Med. 168:140 (March) 1955.

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ARCHIVES OF PEDIATRICS

VOL. 72 SEPTEMBER 1955 No. 9

JOHN FITCH LANDON, M.D., Editor

GARGOYLISM
(Hurler's Syndrome)
REPORT OF THREE CASES

W. F. TOWNSEND-COLES, M.D.
Kitchener School of Medicine,
Khartoum, Sudan.

It seems that no account of gargoylism in Northern Sudanese has been published until now. I have not read of other African native cases though they must have been reported because the disease is said, in textbooks on pediatrics, to have no racial limitations. This paper records briefly three Sudanese gargoyles in whom the characteristics were so marked as to leave no doubt in the diagnosis. All three, who were believed to be unrelated by birth, were typically coarse, ugly dwarfs lacking normal mental development. A blue haze dimmed the corneas, and the livers and spleens were enlarged. Skeletal deformity was general, but was marked in the skulls, which were typically misshapen, and in the backs which were disfigured by kyphosis in the dorsolumbar region. Lateral x-rays of the spine revealed, in all three, the characteristic hook-like anterior deformity of the upper lumbar vertebral bodies. Films of the long limb bones showed thick rectangular shafts and coarse irregular reticulation, instead of the normal shapeliness of outline and structure.

Case 1. A. G. el S., female, aged 3 years. This fretful girl could crawl, but could neither walk nor talk. The head was large with frontal bosses, and the squat nose discharged continuously. (Fig. 1.) All the milk teeth had erupted, but were badly formed. Lateral x-rays of the skull showed a striking deformity of the sella turcica.

(Fig. 3.) The elbows, wrists, knees and ankles were enlarged and were limited in movement to full extension. This patient was accompanied only by her mother, who gave no information about the rest of the family.

Case 2. B. I., female, aged about 2 years. This patient was brought in by her mother who would not give the family history

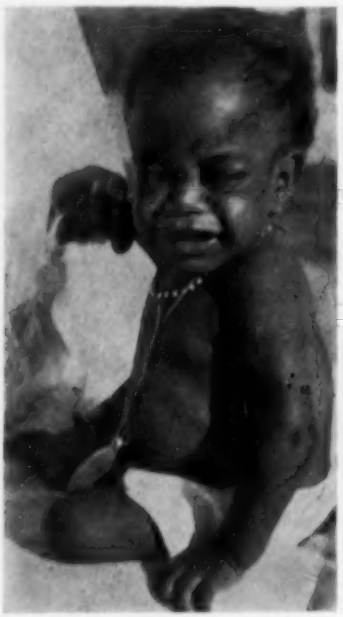

Fig. 1. Patient A. G. el S., aged 3 years.

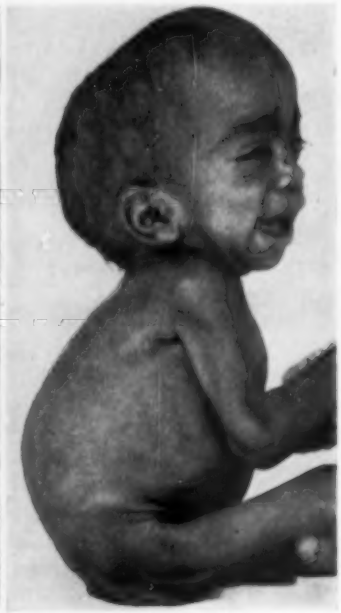

Fig. 2. Patient I. G., aged 2 years.

and refused permission to photograph her child. The typical appearance of gargoylism was associated with typical deformity of the upper lumbar vertebrae seen in lateral x-ray films. Changes in the limb bones were shown radiologically.

Case 3. I. G., male, aged 2 years. (Fig. 2.) This child was the youngest of a family of seven children. All were girls except the oldest and youngest, and all were said to be healthy except this child. Between the births of the first two children, the mother had miscarried, at the eighth month, a male fetus. The parents,

who appeared healthy, were cousins, their fathers being brothers. The patient was still breast fed, and was said by the parents to have had no illness but the head had increased in size rapidly since birth, and double inguinal hernias had appeared at the age of eleven months. At the time of examination, the child's weight was 12 pounds 9 ounces, his height was 27 inches, and the greatest head circumference 19 inches. The child could hold up his head but could not sit up and made no attempt to speak. Very large scalp veins coursed over the deformed skull, and the serrated free mar-

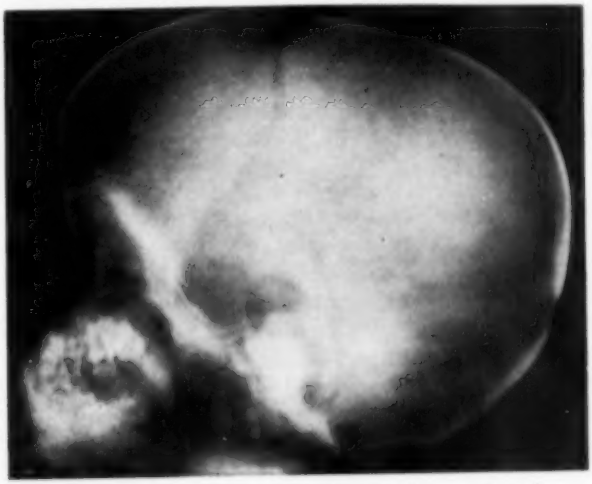

Fig. 3. Showing large sella turcica. Patient A. G. el S.

gins of the four upper and the right lower central incisor teeth were just visible in abnormally wide gums. The palate was low arched. Both testicles were felt lying in the scrotum with bilateral reducible inguinal hernias. Blue pigmented macules of irregular shape and size were situated on the skin of the back. Both plantar reflexes were down-going. The scapulae were placed unduly high on the chest wall, and extension of all the limb joints was limited; x-rays of the spine, and of the rest of the skeleton, showed typical changes.

Manifestations of the disease in these patients conform with those in children of other nationalities recorded in the literature.

HYDROCEPHALUS*

Diagnosis and Treatment

Charles Stabinsky, M.D.
New York.

Hydrocephalus is a persistent and progressive accumulation of cerebrospinal fluid associated with an increase in hydrostatic pressure in the ventricular and/or the subarachnoid system.

Anatomy and Physiology. It is convenient to think of cerebrospinal fluid circulation as two systems: ventricular and subarachnoid. Fluid is produced by the choroid plexus of the ventricular system. As it is formed in the lateral ventricles, it circulates through the foramena of Monro to the third ventricle, then through the aqueduct of Sylvius to the fourth ventricle, and enters the cisterna magna by passing the two lateral foramena of Luschka and the posterior foramen of Magendie. Branches of the cisterna pass into the subarachnoid space over the brain space as spokes of a wheel and into the spinal subarachnoid. The subarachnoid is the site of absorption of the fluid; the cerebral area accounts for 80 per cent and the spinal portion for 20 per cent.

There is no longer any controversy as to the site of production of the fluid. However, dispute still exists as to the mechanism of absorption. The three major hypotheses are: 1. pachionian granulations in the cerebral subarachnoid space; 2. microscopic pores between the subarachnoid space and the longitudinal sinuses; 3. capillary walls of the subarachnoid space. Dandy asserts that the first two theories are untenable because the pachionian granulations are absent in early childhood and the small pores have not been found experimentally. He maintains that cerebrospinal fluid, like other body fluids, is absorbed by the capillary bed according to Starling's Law of the Capillary.

Although cerebrospinal fluid may act as a buffer against cerebral trauma, and carry metabolites to the hemocirculatory system, its primary function is to compensate for space changes within the skull. It has been shown in conditions such as space occupying lesions, e.g., intracranial neoplasm, fluid production decreases, and the ventricles may become minute in size. On the other hand, in maladies where brain tissue is diminished in amount,

*From the New York Medical College, Flower and Fifth Avenue Hospitals, New York.

e.g., cerebral atrophy, fluid production increases and the ventricular system expands to fill the cranial vault.

Cerebrospinal fluid is produced at a greater rate than it is absorbed normally. The rate of production can be augmented by trauma, inflammatory disease, and, as stated above, cerebral atrophy. Rarely, in these conditions, the pial vessels as well as the choroid plexus serve to produce fluid. These diseases are not included in the present discussion because they do not cause hydrocephalus as previously defined.

Classification and Etiology. Hydrocephalus may be caused by (1) increase in production, or (2) inadequate absorption of cerebrospinal fluid. Obstructive phenomena are included with inadequate absorption, because if the fluid does not circulate normally, it will fail to be absorbed.

Clinically, hydrocephalus is classified as (1) internal: excessive fluid or pressure within the ventricular system, or (2) external: excessive fluid or pressure within the subarachnoid system, or (3) combined. The internal type may be further subdivided into (a) communicating: where there is an increased production, or the obstruction is distal to the foramena of Luschka and Magendie, and (b) non-communicating, which is distinguished by a block within the ventricular system.

The most common etiologic factors include: (1) congenital atresia; (2) tumors; and (3) inflammatory sequella. Excessive production of fluid is an extremely rare cause of hydrocephalus. It may occur following acute or subacute intracranial inflammations.

Obstructions of the foramena of Monro are usually due to neoplastic disease, but rarely may be caused by post-inflammatory cicatrix. Such an obstruction results in unilateral dilatation of the lateral ventricle. Ventricular obstructions are caused by neoplasms, such as ependymomas, choroid plexus adenomas and medulloblastomas. These lesions cause dilatation of the ventricular system proximal to the site of obstruction. Congenital atresia is the most common etiology of obstruction of the aqueduct of Sylvius. The sequella is bilaterally equal expansion of the first three ventricles. Post-meningitic adhesions, Arnold-Chiari defect and congenital Bateman's membrane may result in obstruction of the foramena of Luschka and Magendie. Obstruction of

the cisterna magna is most often caused by congenital absence with the meninges opposed to the brain stem, a transverse band across the ventral surface of the brain stem, or post-meningitic scar formation.

Incidence and Prognosis. In one series, 88 living children out of 130,000 live births were found to be hydrocephalic. In infancy, congenitai anomalies are the most common cause of hydrocephalus. However, postinflammatory scars and congenital neoplasms may be etiologically related in this age group. Inflammatory disease is the most frequent cause in early childhood. In children over ten years of age tumors and abcesses are more common causes. It is important to realize that congenital anomalies may not manifest themselves until adult life and may be the cause of hydrocephalus. Hydrocephalus almost never is present at birth unless spina bifida is associated.

Pathology. Hydrocephalus may be associated with other congenital anomalies coincidentally. These include encephalocoele, cerebral anomalies, "forking deformity of the Aqueduct of Sylvius," and Arnold-Chiari deformity. The latter deformity is almost always found when meningomyelocoele exists. The Arnold deformity consists of a tongue-like protrusion of the cerebellum through the foramen magnum. The Chiari defect is an elongated medulla oblongata which obstructs the foramen magnum.

Pathological changes secondary to pressure may consist of cortical thinning, widened sulci and flattened convolutions. The basal ganglia and corpus striatum are peculiarly resistant to pressure damage. The skull bones may be paper-thin, or thicker with irregular inner ridges separating small round or oval defects, covered only by a pericranial membrane. These defects are called "lückenschädel." Irregular projections of brain subtance may project into the markedly dilated ventricles.

External hydrocephalus may rupture into the subdural space. This will force most of the brain tissue to the base of the skull leaving the expanding skull filled with fluid.

Diagnosis. The diagnosis of hydrocephalus is easy to make in the late stages. However, if one waits until the disease is too far advanced, no therapy will be beneficial. Hence, it is necessary to be aware of and recognize the early signs. Once the diagnosis is made, rapid, definitive therapy is indicated.

Some of the early signs which point to the diagnosis are:

1. Fullness or elevation of the fontanelles in a sitting position.

2. Separation of the suture lines up to the age of eight years; between 12 to 15 years of age the skull bones unite and separation no longer occurs.

3. Prominence of the frontal bones with apparent recession of the eyeballs. The weight of the fluid may push the brain toward the base of the skull resulting in interference with total vision as the eyes are forced to the inferior part of the orbit. This can be tested by pulling the lower lids down with a return to normal vision.

4. Widening of the anterior fontanelle.

5. Prominence of the scalp veins.

6. Persistent dullness, irritability and poor weight gain.

7. Circumference of the head exceeds that of the chest.

8. Positive MacEwan's sign—"cracked-pot" resonance on percussion of the skull.

It is interesting to note that mental activity may remain normal, or be slightly impaired with very extensive cortical destruction. Only very late in the progression of the disease do mental aberrations manifest themselves. Other signs include changes in motor function such as spasticity. The only exception to this is when there is obstruction at the base of the ventricular system, e.g., foramena of Luschka and Magendie. In such a block, spasticity occurs early and may be used as a diagnostic point to localize the lesion.

In addition to the aforementioned signs and symptoms, there are some diagnostic tests which may aid in making the diagnosis and help in localizing the lesion. However, it is important to recognize the inherent dangers of certain procedures, to use only those which are essential, to perform as many tests as possible with each procedure, and to use extreme caution.

Included among the tests used are:

1. Skull x-rays: examination of the sutures to note separation or thinning of skull plate.

2. Intracranial tap: The needle is placed first in the subdural space, and if more than 2 cc. of fluid is obtained, the needle is pushed forward into the subarachnoid space, and then into the ventricle. The distance that the needle travels through the brain substance before reaching the ventricle is of prognostic significance and should be noted. The cerebrospinal fluid pressure in the ven-

tricle is normally 50-100 mm. of water. It is considered excessive if it is 150 mm. and dangerous when 300 mm.

Lumbar puncture may be necessary, but it should be done with extreme caution. Medullary choke and instantaneous death may result when a spinal tap is performed with a markedly increased intracranial pressure. If it is essential to perform this test, a small gauge needle (20-22) should be used, and only a few drops of fluid should be removed. The Queckenstadt test should not be routine, because it may precipitate herniation of the medulla oblongata into the foramen magnum.

3. Pneumoencephalogram may show the site of obstruction.

4. Dye studies are used to differentiate the types of hydrocephalus; not to make or confirm the diagnosis. One cc. of neutral phenolsulfophthalein is injected into the spinal subarachnoid space, and twenty minutes later the dye should be found in the ventricles. In two hours 40 per cent of the dye should be excreted in the urine. Indigo carmine, 1-5 cc. of 8 per cent, can be injected into the ventricle. The dye should be found in the opposite ventricle and spinal subarachnoid space immediately. If the above dye tests do not give the normal results, it may be postulated that an obstruction to the flow of fluid exists.

Differential Diagnosis. Macrocephaly may be distinguished by the history of familial occurrence, the width of brain tissue the needle must penetrate before reaching the ventricle, and the normal intraventricular pressure.

Subdural hematoma or hygroma (hydroma) is differentiated by the history of previous trauma and the subdural tap. The fluid in the subdural space in these conditions contains a greater concentration of protein than does cerebrospinal fluid.

Cleidocranial dysostosis is a rare condition which can be determined by x-rays of the skull and clavicles associated with an abnormal range of motion of the upper limbs.

Intracranial neoplasm may show calcified areas on x-ray, or an increased protein in the fluid.

The difficulty in making a definitive diagnosis may be illustrated by the following case history:

CASE REPORT

B.J., a ten-month-old female, entered the hospital with com-

plaints of vomiting, lethargy, trismus and a red, swollen area in the right parietal area. Four weeks prior to admission the patient fell from her stroller and had a convulsive episode without loss of consciousness. The child was well for a few days and then began to regress in development. Vomiting and lethargy set in one week before admission, and trismus two days later. Physical examination on admission revealed a lethargic, poorly nourished child with wide open sutures and fontanelles, positive MacEwan's sign and head circumference of 18½ inches. The differential diagnosis on admission included subdural hematoma, brain tumor, tuberculous meningitis and congenital hydrocephalus.

Skull x-rays showed open fontanelles; subdural tap returned no fluid; lumbar puncture revealed *360* mm. water pressure after no definite papilledema was noted; ventriculogram showed obstruction between the third and fourth ventricles. A neoplasm was considered at this time, but before surgery could be attempted the patient expired.

The post-mortem showed hydrocephalus due to a congenital atresia of the aqueduct of Sylvius with slight incarceration of the temporal lobes below the tentorium.

Therapy. Although no treatment has been consistently successful in curing hydrocephalus, there have been some encouraging results with various modes of therapy. However, there are cases reported which underwent spontaneous remission without recurrence. These include spontaneous rupture of the third ventricle and separation of adhesions. This fact should be considered before radical procedures which have considerable mortality rates are instituted. Therapy in this condition can be divided into (1) conservative, and (2) surgical.

Conservative Treatment should not be continued for more than one month unless marked improvement is evident. Some measures include limitation of fluid intake to as little as 16 ounces per day, diuretics, anti-cholinergics, irradiation of the choroid plexus, postural drainage and repeated intraventricular taps.

Indications for Surgery consist of:

1. Initial and persistent fluid pressure greater than 250 mm. water.
2. Poor clinical results with conservative therapy.
3. Non-communicating hydrocephalus.

Contra-indications to Operative Intervention include:

1. Markedly enlarged head measuring over 50 cm. in diameter.
2. Cerebral cortex which measures less than one centimeter thick.
3. Associated congenital anomalies which prohibit surgery or life expectancy.
4. Poor mental activity.

There have been virtually as many different types of surgical procedures as there are surgeons. Some have met with complete failure and have been abandoned entirely. Others show promise for the future, but require modification. The advantages and disadvantages of each procedure will be mentioned as each procedure is discussed.

Choroid plexus coagulation and extirpation has the advantage in that artificial tubes need not be inserted and will not become obstructed, requiring additional surgery. However, the third and fourth ventricles cannot be reached operatively so there will still be a significant amount of cerebrospinal fluid formed. There is also a considerable increase of post-operative epilepsy with this procedure. Putnam maintains, however, that this technique is very effective and in a recent series of 19 cases he shows 18 survivals, 15 with favorable results and 11 with good mental development.

Third ventriculostomy has several modifications depending on the operator. This procedure has its greatest success in obstruction of the aqueduct of Sylvius in patients over one year of age. Among the important variations of this procedure are: (1) Balkenstitch, where the roof of the third ventricle is opened. The disadvantage lies in the fact that there is marked glial reaction of the overlying brain resulting in closure of the fistula. In a series of 10 infants, 5 had favorable results, and the other 5 died at operation, or shortly thereafter. (2) Puncture of the lateral wall of the ventricle. (3) Puncture of the floor of the ventricle has gained the most favor recently because no brain tissue reaction closes the opening, and the drainage is directly into the cisterna magna, a space large enough for adequate drainage. This technique is valueless if adhesions exist which block the basal cisterna.

Torkildsen's procedure consists of inserting one end of a rubber tube into the occipital horn of the lateral ventricle, drawing the tube subcutaneously, and placing the other end into the cisterna

magna. This method is best used in obstruction of the third ventricle or the aqueduct of Sylvius. However, the rubber tube incites local reaction around it, and, sooner or later, closes off and requires re-operation.

Suboccipital craniectomy with lysis of adhesions is used with good results if post-inflammatory cicatrix is the cause of the hydrocephalus.

Ventriculoperitoneal shunt consists of connecting the lateral ventricle with the peritoneal cavity by polyethylene tubing. The danger of obstruction of the tubing exists. Another disadvantage is the possibility that polyethylene, although almost completely inert, has been shown to be sarcogenic in rats. The possibility of its inducing sarcoma in the human may prohibit its future use.

Lumbar subarachnoid-peritoneal shunt is a procedure used by Scott who introduces polyethylene into the lumber subarachnoid space and passes it subcutaneously to the peritoneal cavity. It would appear that this method would be of use only in communication hydrocephalus, but Scott presents a series of 21 cases, 6 of which are non-communicating, and shows excellent results in 11 of the patients, some of which are the latter type.

Uretero-dural anastomosis is a complicated technique which requires unilateral nephrectomy and anastomosis of a polyethylene tube between the lateral ventricle and the ureter. Major disadvantages include the need for removal of a healthy kidney, the loss of fluid and electrolytes directly into the urine and the upset of the mineral homeostatic mechanism. In a series of 42 cases, 31 showed favorable clinical results.

Ventriculo-mastoidostomy is a procedure which entails the introduction of one end of a polyethylene tube into the lateral ventricle and the other through the mastoid process into the eustachian tube to the pharynx. This technique has a surprisingly low incidence of retrograde infection.

A mechanical modification of the polyethelene anastomotic system is the insertion of an inert metal valve which will prevent retrograde flow of cerebrospinal fluid when intra-abdominal or intra-vesicular pressure is increased.

It can be seen by the number and variety of surgical procedures used in the treatment of hydrocephalus that none is totally ade-

quate. The hope for the future lies in the establishment of an adequate and permanent system of drainage which can be instituted with facility and low surgical mortality.

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COARCTATION OF THE AORTA IN THE INFANT. ITS SURGICAL CORRECTION. (Semaine hop. Paris, 30:2610-2622, June 25, 1954).

Of 15 infants with coarctation of the aorta, 5 were operated on and 10 were not. One of the five died. Eight of the 10 died, and the other 2 had remissions. It appears that surgical treatment is indicated for this condition; the authors recommend it as soon as the diagnosis is made. Two theoretical objections advanced before operation proved to be unfounded. The first was the idea that stenosis of the aortic isthmus in infancy is always a long, hypoplastic segment, as opposed to the diaphragm type seen in adults. The authors did not see any case of the "infantile" type, either at surgery or autopsy. The second objection was that to the long period of aortic clamping necessitated by surgical intervention. It was held that a sufficient collateral circulation would not yet have developed in the pediatric patients, but this was untrue. The four children who survived operation were greatly benefited by it. The one who died had a bronchopulmonary obstruction. This is now considered a contraindication to surgery. Postoperative complications included: abdominal distention; marked diuresis; pulmonary disorders, principally atelectasis from bronchial obstruction; and bulimia. The most important feature in the diagnosis of the coarctation of the infantile aorta is comparative palpation of the radial and femoral arteries. Often no pulse at all is felt in the latter, but, in any case, there is always a significant difference between the two.—*J.A.M.A.*

PERSONAL ORAL HYGIENE FOR CHILDREN*

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Pediatrics is concerned with the health-welfare of young people up to an arbitrary age limit of about twelve years. Although responsibility for health advice, prevention, treatment and management after this period usually devolves upon others, much of the health-welfare beyond childhood is determined largely by what occurs during the pediatric period. This is especially true in relation to dental health.

Lesions Originate Early. A large part of the damaging, and ultimately destructive caries lesions of any given tooth originate within two or three years from the time it erupts into the mouth. Effective preventive measures must be applied before these lesions begin. Treating or filling cavities in a child's teeth does not prevent them. All too often it does not prevent further advancement of such lesions, neither does it prevent the formation of new lesions.

The lesions of the other important disease affecting the teeth—periodontoclasia—the early stages of which consist largely of marginal gingivitis, also originate and are active within two or three years from the time a given tooth erupts into the mouth. Later, some receding of the gums and advancing pyorrhea pockets, mostly between the teeth, are present. These early stage periodontoclasia lesions usually are not recognized as such. Notably their serious significance is not realized by the subject, nor by others. However, they do constitute, in fact, the early stages of progressive, infectious, pathological processes, usually of long duration, which never end until the involved tooth is removed or is finally exfoliated. Treatment, by applications or by operative procedures, of the lesions of this disease, at whatever stage, does not prevent them. Neither does it prevent (although it may retard) further advancement of existing lesions; nor does it prevent formation of new lesions.

Personal Oral Hygiene. Whether new caries or periodontoclasia lesions develop, or whether existing lesions advance further, depends almost entirely upon the personal oral hygiene habits and

*This paper is based upon information derived through studies promoted by facilities, equipment and supplies provided by the School of Medicine, Tulane University of Louisiana.

methods of the individual. The right method not only prevents lesions before they occur, but it also prevents or greatly deters advancement of existing lesions.

The first permanent teeth to erupt—the four first permanent molars—usually come at six to seven years of age. Decay of temporary teeth, and gingivitis about them, should be prevented, but it is absolutely essential, for the dental welfare of the child, and later of the adult, that permanent teeth should be prevented from decaying or having pyorrhea lesions about them. This can be accomplished by application of the correct method of personal oral hygiene—it cannot be accomplished otherwise. Unfortunately, these facts are not as yet well-known.

Most of the permanent teeth erupt and are in place by twelve or thirteen years of age. Most children, at this age, already have incipient caries lesions in several of their teeth, and many have advanced stage lesions (cavities) also. In addition, they have more or less early stage, but surely advancing, periodontoclasia lesions.

Responsibility Rests Upon Parents. This condition results, almost entirely, from previous inadequate, inappropriate and ineffective personal oral hygiene practices. The parents do not know, or have neglected, the method of oral hygiene which must be followed by everyone to maintain cleanliness and dental health. The responsibility rests directly upon the parents and no one else. To permit a normal child to develop even one caries lesion, or one pyorrhea lesion, is a reflection upon the parental care and the personal oral cleanliness practices in the home.

Opportunity For Pediatricians. The purpose of this paper is to direct attention to the very great importance of maintaining effective personal oral hygiene during the pediatric period. It is hoped that many more of those responsible for advice and guidance, relative to the general health of children, will recognize their opportunities in this regard and will actively promote the dental health and well-being of the children in whom they are directly interested. The pediatrician who knows the correct method of personal oral hygiene necessary to prevent the principal diseases of the teeth in children, has a wonderful opportunity to favorably influence the health, welfare and happiness in this regard throughout life, of

those children whose parents depend upon him for advice and guidance. The opportunity for good is very great.

Local Lack of Cleanliness Essential Etiological Condition. The lesions of both caries and periodontoclasia originate only at certain protected areas where bacterial film (called plaque) and decomposing food accumulate, and are retained. They do not originate at locations which are kept clean by functional friction or otherwise. Other factors which may influence the incidence and activity of these diseases are not sufficient, in themselves, to cause lesions at locations which are kept clean. Local lack of cleanliness, therefore, is the essential etiological condition. It is a fundamental fact that "a clean tooth does not decay"; "periodontoclasia does not occur about a clean tooth." It is evident, therefore, that maintenance of a high degree of cleaniness at these vulnerable locations is necessary for prevention of these diseases.

Microscopic Information Necessary. An effective method of cleaning the teeth, and of maintaining the necessary degree of cleanliness, must be based upon the nature of the unclean conditions to be counteracted or minimized, and on the location, sizes, shapes, etc., of the areas and spaces to be cleaned.

These diseases are caused by microscopic organisms. The lesions, at first, are microscopic in extent, and advance microscopically; the tissues involved are composed of microscopic elements and the destructive processes are microchemical. Any method of cleaning the teeth effectively must be based upon correct information as to the local etiological conditions; and this can be derived only through microscopic research and study.

An Effective Method. Thus, and from the published work of others, I have acquired the necessary information, and have designed and described[1] an effective method of personal oral hygiene based upon the essential local etiological conditions in these diseases. By this method, but in no other way now known, the maximum degree of oral cleanliness and dental health can be maintained. This method is different from, and in important particulars is the opposite of, methods generally followed. The teeth must be cleaned right with the right kind of both toothbrush and dental floss every night before retiring. The considerations upon which the method is based and the method itself have been described in

detail in the paper referred to above, and in other publications. They need not be repeated here.

Each Person Must Be Taught. Oral cleanliness is largely an individual and personal matter. Each person must be taught the right method by someone who knows and understands the conditions, and knows how to teach it to others. He would be following the method himself. It would be a rule in his own home that no one retires at night with retained decomposing food material and other conditions of uncleanliness about his teeth.

Only One Effective Method Now Known. There is only one effective method of personal oral hygiene now known. The almost universal prevalence and ultimate destructiveness of both caries and periodontoclasia confirm the ineffectiveness of all other methods in general use heretofore. Although some changes or improvements may be indicated by future research and experience, it is certain that, for the present at least, this exact method must be known and followed faithfully, to adequately maintain oral cleanliness and dental health. In the light of the facts which we now know as to the essential local etiological conditions in these diseases, it is obvious that any neglect or departure from this exact method would lessen the effectiveness to the full extent of such neglect or departure. Under these circumstances, each person who wishes to maintain his or her own oral cleanliness and dental health, and each one who has responsibility for the oral cleanliness and dental health of young children must be taught and then follow and apply this method.

The need is for trained teachers who know and can successfully teach the method to others. They must know and understand the local etiological conditions in these diseases and how they can be controlled. After short periods of instruction and experience, practicing dentists are able successfully to teach this method of personal oral hygiene to a good proportion of their patients in connection with their treatment and restorative services. The need is for many more to have the necessary information and to be prepared to teach this effective method to those who wish to follow it.

Application To Children. Parents who know and follow this method are then, and only then, able to bring the full benefit of it to their children. It should be a rule in the home that preparation

for retirement at night must include cleaning the teeth properly. In most instances this can be done well enough with the toothbrush followed by rinsing to protect the deciduous teeth from caries lesions. From the time the child is two or three years of age his teeth should be cleaned properly by the parent. The young child cannot brush his teeth effectively. It must be done for him by someone who knows how it should be done.

It is vital that the occlusal surfaces of the grinder teeth be thoroughly cleansed. This is accomplished by applying the bristles of the brush directly and firmly to these surfaces and making short back and forth movements so as to dig out and remove microscopic (and sometimes macroscopic) material lodged and retained in the pit and fissure depressions which are present on the occlusal surfaces. The teeth in each quadrant must be cleaned in this way.

At the same time the buccal, labial and lingual surfaces of all the teeth should be brushed. Here, too, the bristles of the right kind of brush should be directed into the sulci between the teeth. The same short stroke motion is most effective in digging out material from these locations and cleaning the surfaces of the teeth. Brushing should be followed by rinsing the mouth. By perseverence, the child can be taught, in time, to rinse his teeth well after brushing. Vigorous rinsing is an important part of the oral hygiene procedure for all people.

After the first permanent molars have erupted and attained their normal occlusal level then the right kind of dental floss must be passed between them and the contacting deciduous molars. It is only necessary to pass the floss through the contact area and back out. This insures against proximal caries lesions. It is not necessary for the young child to carry the floss into the gingival crevices as must be done by older children and by all adults to clean the teeth within the gingival crevices. As other permanent teeth come in, the floss must be passed between them and any other teeth with which they are in contact.

Parent Must Clean Child's Teeth For Him Until He Can Do It Himself. The parent must clean the child's teeth for him until such time as he himself can do it effectively. At what age and when any given child can learn and be allowed to take over this procedure will vary considerably, depending largely upon his ability and will-

ingness to acquire the necessary manual dexterity, and upon the parents' ability, and patience, to train him.

SUMMARY

Many of the lesions of the two principal diseases of the teeth originate during early childhood.

Local lack of cleanliness and poor oral hygiene are essential for the origination and advancement of the lesions. Prevention or minimizing of these conditions is necessary to prevent the diseases.

An effective method of personal oral hygiene is now known, based upon accurate information as to the microscopic conditions at the vulnerable locations. Practical application of this method to young children is discussed.

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DIABETES MELLITUS IN TWINS, WITH A REPORT OF FOUR CASES. (Glasgow M. J., 36:15-19, Jan. 1955). Two sets of diabetic twins have been under the author's care for from two to seven years. In the first pair (male, uniovular), the disease developed in the older twin at the age of 28 years and in the younger three and a half years later. In the second pair (female, binovular), diabetes first appeared in the older sister at the age of 54 years and in the younger six months later. All four are maintained in good health with insulin. No history of diabetes could be found in the two families, but the disease could have been present in the grandparents. If, as many authorities believe, diabetes mellitus is transmitted by a recessive gene according to Mendelian law, then skipping of a generation is to be expected. The evidence provided by the cases of diabetes in twins reported in the literature shows that heredity is at least one factor. Both members of a pair of uniovular twins are about five times more frequently affected than are both binovular twins. Almost without exception in uniovular twins, both are said to suffer from diabetes after the age of 43. When only one has the disease, his partner is likely to have an abnormal glucose-tolerance test curve. Both partners of the first-affected twins in this series refused testing.—*J.A.M.A.*

PEDIATRICS AT THE TURN OF THE CENTURY

From time to time the Archives, which was the first Children's Journal in the English language, will reprint contributions by the pioneers of the specialty over fifty years ago. It is believed that our readers will be interested in reviewing such early pediatric thought.

DIAGNOSIS OF LATE HEREDITARY SYPHILIS IN THE SCHOOL CHILD*

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It is my purpose tonight to present to you the subject of the diagnosis of heredo-syphilis, not only from the viewpoint of the practising physician, but also from that of the medical examiner of school children. To this end I shall consider chiefly such phenomena of heredo-syphilis as are peculiarly frequent at school age, and are discernible during the routine physical examination which the medical examiner makes at school.

Late heredo-syphilis—the only form of heredo-syphilis which can occur in school childen—is that phase of the affection which appears after the third year of life. On the other hand, the phenomena of heredo-syphilis which appear before the third year may well be designated as "early" hereditary syphilis.

For a long time, down to a comparatively recent period, it was held that the early signs of the disease were the only manifestations of specific hereditary infection that were to be feared, and that treatment instituted in infancy eradicated the taint, so that no further consequences of it were to be expected late in life.

The multiform signs which heredo-syphilitic children exhibited in later years were commonly held to be "scrofulous" in origin, and it was not until the brilliant researches of Hutchinson, Fournier, Parrot and others had appeared, that physicians came to realize that syphilis acquired in utero may last through youth

*Read before the Society of the Medical Inspectors of the City of New York, Tuesday, June 5, 1906.
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Reprinted from ARCHIVES OF PEDIATRICS, 23: 914-924, December 1906

to adult age, just as syphilis acquired at twenty may persist to the Seventh Age of Man. Modern research has, furthermore, demonstrated that an infant treated most thoroughly for heredo-syphilis may show no evidence of the disease until he reaches school age, or even adolescence or adult life, and that after an interval of many years such a person may run the gauntlet of "late" heredo-syphilis in all its phases.

In the school child, therefore, we may have three possibilities as to the presence of heredo-syphilitic symptoms: we may, first of all, have traces of an *early* heredo-syphilis, as, for example, scars of old lesions on the skin or about the mouth or nostrils; we may, secondly, have signs of a *late* heredo-syphilis which, however, is no longer active; and finally, we may have symptoms of an *active* heredo-syphilitic process.

That heredo-syphilis exists in the school children of this city is a fact which cannot be gainsaid—a fact that does not evoke astonishment. When one remembers the motley sources of our population and the frequency of syphilitic infection among some of the races that contribute to the total of New York's citizenship, the comparative infrequency with which the school examiner finds evidence of heredo-syphilis among these children may well cause wonder.

In partial explanation of the apparent rarity of heredo-syphilis in our public schools, I may offer two factors: first, the circumstance that the signs of heredo-syphilis are not usually so glaring that he who runs may read, but are apt to escape notice or be mistaken for something else unless one knows them well and knows where and how to look for them; secondly, that a scientific diagnosis of heredo-syphilis cannot be made without a positive family history of luetic infection, even though the classical signs of the hereditary affection be present, and that unfortunately such a family history is almost impossible to obtain for the school inspector, unless he secures the cooperation of the family physician.

So far as my personal observations go, I have been able to find but 12 cases in which hereditary syphilis could be diagnosed from the presumptive evidence of physical signs, among about 2,500 school children whom I examined during the past year. There were also a number of children among these in whom a

suspicion of heredo-syphilis was evoked by one or more physical features, but in whom the evidence was not sufficient for a diagnosis. Owing to uncontrollable circumstances, I could not in any of these cases get a family history, but in a number of instances I have had the opportunity to examine several children in the same family, and it was thus that I was able to strengthen my evidence and to make the diagnoses more positive. I shall take occasion to speak of a few of my more interesting cases in discussing the various physical signs of heredo-syphilis.

1. *Age. General Appearance.* Late heredo-syphilis presents itself most frequently at about twelve years of age. My own cases range from six to fourteen in age, and 5 of them were in boys while 7 of the patients were girls. In general appearance, heredo-syphilitic children present many features which, while not characteristic enough to be pathognomonic, are warning signals for further examination. The children affected are usually delicate, sickly-looking, ill-nourished, with flabby muscles and with a typical pallor which has often a grayish tinge that has been made classical by Hutchinson. This author goes so far as to consider the peculiar grayish skin of heredo-syphilitics as one of the most characteristic outward signs of the affection. The appearance of heredo-syphilitic children must be carefully differentiated from that of scrofulous children, children with what is known as the "fine type" of tuberculous individuals, with delicate pale or pink cheeks, finely chiseled features, thin lips, fine lower jaw, bright eyes and long eyelashes.

Physical, and as we shall see later, mental, development is regularly retarded by heredo-syphilis, at times to such a degree as to present persistent *infantilism* at an advanced age. These children begin to walk and to talk late; when seen in the lowest grade of the school they cannot do the simplest tasks that others of their age perform with ease. In boys the physical signs of puberty appear late; quite the reverse of what is seen in backward children with signs of degeneration, whose puberty is precocious, as a rule. In girls the menstrual function, the breasts, and the other secondary signs of sex appear severel years later than normally, at seventeen instead of at fourteen or fifteen. Retarded development, indeed, is seen at all ages in heredo-syphilitics. Girls of fourteen may look like children of six (Fournier) and women of forty like girls of

sixteen (Lancereaux). In one of the cases which I saw in my work as examining physician for the St. John's Guild Hospitals, a little girl of two and a half years could not possibly be given more than nine months from her appearance and size:

CASE I. Retarded development: Heredo-syphilis: Rickets. Stanislaus M., two and a half years old. Parents, Polish; father, a brassworker by trade, three years in this country. The family history is not definitely negative. This child looks like a baby nine months old so far as size and development are concerned. When he opens his mouth, however, it appears that his teeth are well developed for his age. He has a bulging forehead, a hydrocephalic head, a moderately depressed nose, scars at the corners of the mouth and there is the history of a series of eruptions during the first months of his life. He presents, moreover, all the characteristics of advanced rickets, including beaded ribs, a rachitic spine, bowlegs, atrophy of the long bones. The superficial glands in the neck and groin are enlarged. Here, then, is an example of marked under-development due to the combined factors of rickets and heredo-syphilis. As yet I have not been able to trace the further history of this child, but it would be interesting to see in how far mercurial treatment might affect his growth.

2. *Deformities of the Skeleton.* The skull is frequently the seat of deformities in heredo-syphilis. A bulging forehead, often with two lateral bosses over the brows, is frequently seen. Parietal, occipital and other bosses are also sometimes seen, as well as asymmetric heads, and skulls with transverse enlargement. *Hydrocephalus*, according to Fournier, is very common among heredo-syphilitics, and occurred in three of the twelve school children. Hydrocephalus should indeed always lead us to an investigation as to the possible presence of heredo-syphilitic signs.

Deformities of the Chest, the Spine and the Extremities are very common in heredo-syphilitic children. The deformities characteristic of rickets, such as pigeon-chest, beaded ribs, bowlegs, etc., are often seen; in fact, you will remember that Parrot's theory of rickets is, that rachitic deformities are due to syphilitic taint. The origin of this theory lay in the fact that rickets was so often found in heredo-syphilis. The accepted view is, however, that heredo-syphilis simply predisposes to rickets. Much more charac-

teristic deformities are seen in the hypertrophies of the long bones, especially at their ends, as at the elbows, the knees or the ankles, where we find enlargements, exostoses and a variety of other deformities. The most typical deformity of the bones, however, is *the thickening of the crest of the tibia*, which is curved like a sabre from behind forward. Hutchinson insists that we should never fail to feel the tibia in a suspect of heredo-syphilis. Fournier calls it the "revealing bone" (*"os révélateur"*).

3. *Lesions of the Skin.* In children at school age hereditary syphilis does not tend to affect the skin, save in two forms: *syphilitic lupus and gummatous ulceration*, both of which are comparatively rare. The former resembles in most respects the lesion known as syphilitic lupus in adults, and consists of an often multiple, rather extensive infiltration, with crescentic outlines and a surface of brownish or coppery hue. It heals rapidly under the influence of mercury and leaves thin, flexible white scars. The gummatous lesions are especially common on the legs in the anterior tibial region.

Scars of previous lesions are much more common and should be looked for in all cases. First in frequency and importance come the radiating scars *at the corners of the mouth* due to healed rhagades. These are faint tapering whitish streaks, horizontal or oblique, usually multiple, on either side of the mouth. Other scars may be seen on the body, especially on the buttocks, as traces of eruptions in infancy, but these are inaccessible to the examiner of school children and need not be described.

4. *Enlarged Glands.* Enlarged lymph nodes are, of course, in no way characteristic of heredo-syphilis; yet they are almost always present in these children. They are usually multiple indurated, discrete and persistent enlargements of the cervical, axillary or inguinal lymph nodes, which reach their acme and remain stationary for a long time, but do not tend to suppurate.

5. *Lesions in the Nose and Throat.* Both these cavities should be carefully scrutinized, as they reveal a number of characteristic lesions in heredo-syphilis. The appearance of the nose is often suggestive. We all know the *depressed nose* of heredo-syphilis which even laymen nowadays recognize. This depressed nose may be one of two types. The first is what may be styled the necrotic

type, secondary to a crumbling of the nasal bones, especially of the nasal septum, due to caries. This type is manifested either by a depression of the root of the nose, the point of the organ being tipped up, or else by a sulcus obliquely dividing the lower from the upper two-thirds of the nose, resulting from an invagination of the lower portion into the upper. The second type of nasal depression in heredo-syphilis is more commonly met with in young children, and is the hereditary or congenital form, not due to necrosis. It is manifested simply by a depression or a flattening of the upper part of the nose without any effect on the lower segment.

Examining the nostrils, we find often a foul-smelling, ichorous discharge of sero-pus which may be streaked with blood, and we recognize the overpowering odor of ozena—a common lesion in heredo-syphilis, though not pathognomonic of it. It precedes more or less extensive caries of the septum or of other parts of the nose. In school children a chronic "cold in the head," unless due to adenoids or nasal obstruction, is apt to be syphilitic in origin, and it is this apparently trifling affection that forebodes serious involvment of the nasal bones, destruction and deformity. Necrotic processes extending from the nose also may cause perforations of the palate.

In the throat we may find scars of previous gummatous lesions, adhesions and deformities of the palate due to these scars, or an active gummatous process. The palate is then dark red, swollen, indurated and deformed, or the ulceration is seen in its further stages, finally terminating in the clean-cut, oval or oblong perforation so characteristic of heredo-syphilis. The pillars and tonsils and the tongue or the buccal membrane are far less frequently affected. The change of voice which appears with perforations of the palate is familiar enough.

6. *Hutchinson's Triad.* (*a*) *Lesions of the Teeth.* I now come to the most characteristic set of lesions which are seen in heredo-syphilis in the school child, namely to that group of manifestations known as Hutchinson's triad, after the famous English syphilographer who first described them in their true diagnostic light. The signs heretofore spoken of, with the exception perhaps of the nasal and pharyngeal lesions, are merely corroborative signs leading

to a diagnosis, but the triad of lesions to be mentioned now constitutes, when present, presumptive evidence of heredo-syphilis. Indeed, in the face of these signs, a denial of syphilitic infection on the part of the parents must be looked upon most skeptically. These three signs are: Deformities of the teeth with special characteristics; lesions of the cornea, the iris, and sometimes of other parts of the eye; and disturbances of hearing.

Dental Deformities. The anomalies of the teeth found in heredo-syphilis are but an expression of the tendency which the disease has to retard and distort growth and development. Dentition in infancy is retarded usually in heredo-syphilis. Thus Demarquay found no teeth in a child four years old. The lateral incisors and the canines are also sometimes retarded (Lancereaux). This rule has exceptions, however, just as it has in rickets.

Deformities of the *second set of teeth* interest us particularly, as they are more characteristic. A variety of types is seen, including eroded surfaces, grooved teeth, teeth of abnormally small size, abnormally situated, etc. The first molars have a tendency to atrophy in heredo-syphilis, as in all conditions of physical and mental degeneracy. In several of my cases I have found deformities of the teeth, and atrophy of the first molars alone or of some of the other teeth. Caries is very frequent in these children, as the teeth are most prone to decay, and to break off.

The most characteristic deformity, however, that which alone is diagnostic, is the semilunar erosion of the incisors, especially the middle upper incisors of the second or permanent set, known as *Hutchinson's teeth*. These must be familiar to you, as they are figured in most text-books and are easily recognized when typical. Unfortunately, they are by no means always present in heredo-syphilis, but when they do occur, the diagnosis can be made almost with certainty. The free border of these teeth is concave, semilunar, with a beveled edge anteriorly. This beveled edge is very characteristic and should always be looked for. The corners are rounded, instead of sharply passing to the sides of the teeth. The incisors thus affected usually lean toward each other, and are short and low. Although these teeth look as though they have been worn down with use, this is not the case, for they are thus deformed in the dental follicle before they appear on the surface of

the gum. After many years, say at thirty, the deformity is no longer seen and the teeth merely appear as short stumps.

Finally, before dismissing the subject of dental deformities, we may speak of a sign dwelt upon considerably by Fournier, which is said to be very characteristic, if not pathognomonic, namely, *a horizontal white streak* appearing on the surface of the incisor teeth which may or may not be deformed. Fournier has never failed to find a syphilitic family history in children who presented such streaks. We have heard of the "yellow streak," but it seems we must also be on our guard against the "white streak."

CASE II. Hutchinson's Teeth: Hydrocephalus: Depressed Nose, etc. Josephine O., ten years old, of Bohemian parentage, in Class 2A, in a public school, i.e., about two and a half years behind the grade in which girls of her age belong. The family history could not be obtained. This child presents a good example of Hutchinson's deformity on the middle upper and to a lesser degree on the lower incisors. She has also the characteristic ashy pallor, the hydrocephalic head, and the depressed nose of heredo-syphilis, as well as scars at the corners of the lips. No active manifestations of heredo-syphilis have appeared during the past year and a half during which I have had her under observation.

CASE III. Louise O., sister of the above patient, a girl twelve years old, who is now in the 3A grade, shows no Hutchinson's teeth, but a well-marked atrophy of the first molars and caries of the upper molars. Her lower canines are very small, rudimentary. She has also a bulging forehead, though less marked than her sister, and a depressed nose which is less conspicuous. The interesting feature in this girl, however, is a well-marked mitral insufficiency manifested by a classical murmur. No history of scarlatina or of any other cause of the cardiac trouble can be elicited. There may be here a possible connection between the hereditary taint and the heart lesion, although, of course, this is merely a theoretical supposition.

(*b*) The eye lesions which form part of Hutchinson's triad were described as early as 1859 by the observer, and consist of keratitis, iritis, choroiditis and retinitis. *Keratitis* is the most frequent of these lesions. It comes on insidiously, without pain, without or with very slight inflammatory lesions, and appears first as a

series of small grayish points which look like dust or ground glass on the cornea. Gradually the opacity grows more marked, and an inflammatory reaction sets in. Circumcorneal injection, vascularization of the cornea follow, the cornea becoming salmon pink, then cerise-colored, then dark red; the patient feels pain in the eye, has photophobia and blepharospasm. In the course of months the opacity may be absorbed, or it may grow dense, white and remain as a leukoma.

I have had the opportunity of watching the development of a heredo-syphilitic keratitis in a school child whose history follows:

CASE IV. The patient is a little girl aged seven years, of Italian parentage, who from the age of four months to her sixth year had been subject to convulsions. She presents the bulging forehead and the depressed nose in moderate degree, shows faint scars at the corners of the mouth, and is poorly developed physically and mentally. Two of her sisters had the same affection of the eye. One of them is now entirely free from eye lesions and is under my observation, a girl of twelve. At the age of five or six this child was almost blind, the mother says, with a cloud over her eyes which later disappeared. The other sister was a child of three years, who died last March after an illness of two years, manifested chiefly by convulsions at frequent intervals. This little child was almost blind and showed the same cloudy corneae which gradually grew white. I did not see this child, but obtained her history from the family in the words which I have used.

Here, then, we have a family with three children, all of whom at one time have had syphilitic keratitis. The convulsive seizures from which these children suffered are, as we shall see later, also signs of heredo-syphilis as they very commonly occur in this condition. D. R., the girl who now has the keratitis, presents in her right eye the process in the active stage, while in her left eye she has a white spot in the center of the cornea, as a vestige of a keratitis now passed away. I have watched this child for a number of months and persistent efforts to get her under treatment have failed thus far. In such a case it is almost impossible to make some parents believe that anything can be done for the eyes. They are resigned to have the child go through the disease as the elder daughter did.

Iritis is less frequent in heredo-syphilis, but none the less characteristic. I shall not go into its description in detail. There are two principal types: the one resembles in all respects ordinary iritis, save that it is slow in development and mild as to inflammatory signs, and often results in an exudate in the region of the pupil. The other is the gummatous type, which is seen also in adults. Small yellowish brown nodules appear at or near the margin of the iris, which disappear on treatment. Choroiditis and lesions in the eye fundus are accessible only to ophthalmoscopic examination.

(*c*) *Affections of the Ear* in heredo-syphilis may be either secondary to an otitis media, and in this case do not differ from the ordinary chronic middle ear inflammations, or they may be primary, syphilitic from the start. In this case there may be syphilitic otitis media which comes on suddenly, with a discharge from the ear through a tympanic perforation, without any pain or fever. This is followed by deafness unless promptly and efficiently treated. In other cases, and these are by far the most characteristic of heredo-syphilis, there is a sudden rapid, very complete deafness, first in one ear then in the other, without any lesions whatever in the drum or the middle ear. This is central syphilitic deafness due to an affection of the auditory centre in the floor of the fourth ventricle. It is incurable, persistent, absolutely painless, and is accompanied by subjective noises in the ears which cannot be relieved.

7. Nervous and Mental Affections. It remains for us to consider the numerous nervous and mental disorders of heredo-syphilis, to which, however, we shall devote but a few words. Epilepsy is very common in heredo-syphilitics, and every epileptic child should be examined for the signs of hereditary taint. Convulsions in infancy often precede this state, and are also often seen in children with hereditary syphilis. Cerebral heredo-syphilis is, fortunately, not very frequent, for it is by far the most terrible phase of this disease. When it is present, it begins with epileptic seizures or with headaches then goes on to vertigo, partial paralyses, hemiplegias, sensory troubles, etc., just as in acquired cerebral syphilis. Less common syphilitic nervous diseases in children are heredo-syphilitic tabes and disseminated sclerosis.

The mentality of heredo-syphilitics, as has already been hinted at, is very often deficient. We may look for our cases of heredo-syphilis safely among the backward children. In the more pronounced cases they are even imbeciles and idiots, and no history of such mentally deficient children is complete without an investigation as to hereditary syphilis.

SUMMARY AND CONCLUSIONS

1. Among 2,500 school children who were subjected to a thorough physical examination, the author found but twelve in whom physical signs justified a presumptive diagnosis of heredo-syphilis. Owing to the peculair conditions of the investigation no family histories could be obtained.

2. The children examined ranged from six to sixteen years, and about equal numbers of boys and girls were studied. The heredo-syphilitic children ranged from six to fourteen years of age. Five were boys, and seven girls. All save one were deficient mentally; eight children showed Hutchinson's teeth; four showed the remains of syphilitic eye lesions and one showed an active keratitis. Three of the twelve cases had hydrocephalic heads. All showed retarded physical development, and seven of the total number showed associated rickety changes in the bones. Nine showed scars about the mouth; all had enlarged glands; two had lesions of the nose and none in the throat. Chorea was present in two.

3. While heredo-syphilis does not seem to be common in the children of the New York public schools, it constitutes an affection which must be looked for by the school examiner; and if sufficiently characteristic signs are found, he is justified in calling the family physician's attention to these signs as indicating a possible specific taint. The duty of the family physician in such cases is obvious.

DEPARTMENT OF ABSTRACTS

BUCH, C.: EXPOSURE TO VIRUS DISEASES IN EARLY PREGNANCY AND CONGENITAL MALFORMATIONS. (Canadian Medical Association Journal, *72*:744, May 15, 1955).

The infant death rate in Canada from congenital malformations has been found to fluctuate seasonally and from year to year over the period from January 1944 to July 1951. There is no convincing evidence that these fluctuations are associated with variations in maternal exposure to German measles, measles, mumps, chickenpox, influenza or poliomyelitis in the first trimester of pregnancy.

AUTHOR'S SUMMARY.

STOLLERMAN, G. H.; RUSOFF, J. H. AND HIRSCHFELD, I.: PROPHYLAXIS AGAINST GROUP A STREPTOCOCCI IN RHEUMATIC FEVER. THE USE OF SINGLE MONTHLY INJECTIONS OF BENZATHINE PENICILLIN G. (New England Journal Medicine, *252*:787, May 12, 1955).

No recurrences of rheumatic fever were observed among 145 patients with rheumatic fever who received monthly injections of 1,200,000 units of benzathine penicillin during a 2-year-period. Evidence of infection was noted in but 4 of these patients during the period of study. Only 1 of the 4 had a clinically apparent pharyngitis associated with a positive culture for Group A streptococci and a rise in antistreptolysin O titer. The remainder had either a rise in antibody or a positive culture for Group A streptococci as sole evidence of infection. Cardiac lesions progressed in 3 patients who had rheumatic heart disease at the beginning of the study. Those who did not have apparent rheumatic heart disease at the beginning of the study did not have cardiac lesions subsequently. Reactions considered to be due to penicillin hypersensitivity occurred in 5 of 410 patients who received an average of 12 monthly injections per patient. Single monthly injections of 1,200,000 U. of benzathine penicillin confer a high degree of continuous protection against infection with Group A streptococci and afford a reliable means of protecting the patient against recurrences of rheumatic fever.

AUTHORS' SUMMARY.

KNELLER, L. A.; UHL, H. S. M. AND BREM, J.: SUCCESSFUL CALCIUM DISODIUM ETHYLENE DIAMINE TETRA-ACETATE TREATMENT OF LEAD POISONING IN AN INFANT. (New England Journal Medicine, 252:338, March 3, 1955).

Lead intoxication is a prevalent and important disease with a high morbidity, especially in children. A case of lead intoxication and encephalopathy in a 20-month-old child is presented, together with clinical and laboratory findings necessary to establish a diagnosis. A brief outline of the historical treatment of lead poisoning and a commentary on the diagnostic procedures are given. The use of a new therapeutic agent, EDTA (ethylene diamine tetra-acetic acid), or "Versene" is discussed. A modified method of administration of the calcium disodium salt useful in pediatrics and general medicine is described. Calcium EDTA is effective and easily administered. It is recommended as the drug of choice in lead intoxication. AUTHORS' SUMMARY.

SIEGEL, M.; GREENBERG, M. AND STONE, P.: RISK OF PARALYTIC AND NONPARALYTIC FORMS OF POLIOMYELITIS TO HOUSEHOLD CONTACTS IN NONEPIDEMIC YEARS. (New England Journal Medicine, 252:752, May 5, 1955).

The probability of clinical infection among household members after the onset of poliomyelitis in the family was studied in New York City during the nonepidemic period of 1950-1953, inclusive. The data examined were obtained from an investigation of 3,028 household units in which one or more cases of poliomyelitis occurred during the 4-year-period of observation. Multiple cases were reported in 2.7 per cent of these households. The probability that paralytic infection would develop in household members after the onset of a case in the family was greater when the initial case was paralytic than when it was nonparalytic. On the other hand, the risk of subsequent nonparalytic infection seemed to be greater when the initial case was nonparalytic. These differences were most striking among household members under 15 years of age. The attack rate among household members after the occurrence of the first case compared to that observed in the general population was greater in the nonepidemic years of 1950-53 than in the epidemic of 1949. AUTHORS' SUMMARY.

EDMUNDS, P. N.; ELIAS-JONES, T. F.; FORFAR, J. O. AND BALF, C. L.: PATHOGENIC STAPHYLOCOCCI IN THE ENVIRONMENT OF THE NEWBORN INFANT. (British Medical Journal, 4920:990, April 23, 1955).

Over 51 per cent of the members of the nursing, medical and domestic staff in three Edinburgh maternity units were found to be carrying Staph. pyogenes in their anterior nares. Of these staphylococci, 58 per cent were completely resistant to penicillin and nearly 20 per cent were resistant to sulphonamide. Less than 10 per cent were resistant to chloramphenicol, chlortetracycline and streptomycin. The staphylococcal carriage rate of the mothers admitted to, and babies delivered in, these units were determined. The mother's vaginal carriage rate was 3.5 per cent and their nasal carriage rate was 28.7 per cent. Of these, only the vaginal carriage rate rose during an 8-day-period. The babies' eye carriage rate on the fourth day after delivery was 35.3 per cent and the umbilical carriage rate on the eighth day was 40.1 per cent. The corresponding carriage rate for babies born at home were 5 per cent and 8.6 per cent. As between these different maternity units of different type, there was little difference in the mothers, babies, and staff carriage rates. There was a much higher incidence of mothers, and among week-old babies born in hospital than among those born at home. Individual hospitals showed little difference in antibiotic-resistance rates. A relatively high frequency of mild skin and conjunctival sepsis contrasted with a complete absence of serious infection in this series. The great majority of infants acquired their staphylococci from sources other than their mothers, chiefly from the nursing staff and from other babies. Despite the fact that there was a high carriage rate among the nurses attending babies born at home the carriage rate on these babies remained low. Where infants carried staphylococci on two different site (eye and umbilicus), in approximately half the cases the staphylococci had different drug-sensitivity patterns.

AUTHORS' SUMMARY.

TREATMENT OF ACUTE RHEUMATIC FEVER IN CHILDREN. A CO-OPERATIVE CLINICAL TRIAL OF A.C.T.H., CORTISONE, AND ASPIRIN. A JOINT REPORT BY THE RHEUMATIC FEVER WORKING PARTY OF THE MEDICAL RESEARCH COUNCIL OF GREAT BRITAIN AND THE SUBCOMMITTEE OF PRINCIPAL INVESTIGATORS OF THE AMERICAN COUNCIL ON RHEUMATIC FEVER AND CONGENITAL HEART DISEASE, AMERICAN HEART ASSOCIATION. (British Medical Journal, 4913:555, March 5, 1955).

Six centres in the United Kingdom, 5 in the United States and 1 in Canada have collaborated in a trial in the relative merits of A.C.T.H., cortisone and aspirin in the treatment of acute rheumatic fever and the prevention of rheumatic heart disease. The present report relates to children under the age of 16 and compares the effects of the three drugs on the acute course of the disease and on the persistence and development of rheumatic heart disease through one subsequent year. The records of 497 patients are presented (240 U.K. and 257 U.S., including the Canadian centre in the latter). Each case met specified diagnostic criteria on admission to the trial and was allocated at random to treatment with one of the three durgs. Each treatment was given for 6 weeks according to a defined schedule, and detailed observations were continued for a further 3 weeks. Follow-up examinations were made at specified times after these 9 weeks, and the present report extends to the examination made one year later, i.e., 61 weeks, from the start of the treatment. The study was designed to ensure a balance of cases in the 3 treatment groups for each centre, for the duration of illness at start of treatment, and for the time of year when cases were admitted. Random allocation of cases within this balanced design was relied upon to secure a reasonably equal distribution of cases according to age, sex, and severity and frequency of manifestations of disease. This design permits many comparisons of the total groups on each treatment. In 51 per cent of the patients, treatment was begun within 14 days of the onset of the attack; in nearly two-thirds there was no history of a previous attack or evidence of pre-existing rheumatic heart disease. The treatments were therefore tested in patients of whom a large proportion were still in the early stages of the disease and had no established heart disease. The three randomly constructed groups

on A.C.T.H. (162 cases), cortisone (167 cases) and aspirin (168 cases) were notably alike in most respects at the start of the trial. The results of the treatment were measured in relation to separate manfestations of the disease, namely, temperature, pulse rate during sleep, E.S.R., joint involvment, chorea, erythema marginatum, nodules, and such aspects of the status of the heart as heart size, A-V conduction time, murmurs, and, in particular, those indicative of serious illness, congestive failure, and pericarditis. There was no evidence that any of these agents resulted in uniform termination of the disease, and on all treatments some patients developed fresh manifestations during treatment. Treatment with either of the hormones resulted in more prompt control of certain acute manifestations, but this more rapid disappearance was balanced by a greater tendency for the acute manifestations to reappear for a limited period upon cessation of treatment. Treatment with the hormones also led to more rapid disappearance of nodules and soft apical systolic murmurs. At the end of one year there was no significant difference between the three treatment groups in the status of the heart. During the period of treatment, observation, and one year of follow-up, there were only six deaths among 497 cases, under the age of 16, admitted to the study.

AUTHORS' SUMMARY.

SALAZAR-MALLÉN, M. AND RULFO, J.: ON SOME FEATURES OF RHEUMATIC FEVER AND RHEUMATIC HEART DISEASE AS SEEN IN THE NATIONAL CARDIOLOGICAL INSTITUTE OF MEXICO. (Annals of Internal Medicine, 42:607, March 1955).

The following conclusions have been reached as the result of a thorough review of the records of the National Cardiological Institute since 1944: (A) Rheumatic heart disease is widespread in the Mexican Republic. Considering the rheumatic index, its prevalence is comparable to that found in temperate climates in such countries as England and the United States. (B) The presence of a large Indian component in the Mexican population is not considered significant in relation to rheumatic predisposition. If there are more rheumatics among the economically needy classes (having a greater indigenous component), the phenomenon should be attributed to the economic-social factor and not to the racial

(genetic) traits. (C) It has been substantially proved that the female sex is more susceptible to rheumatism. This is noted during the prepubertal age as well as during the second and third decades of the feminine life, hence suggesting a greater hypophysiocortical instability in women, especially during their more active genital period. (D) Climate has a decisive influence on the development of rheumatism. When the variables constituted by the different genetic or economic factors are discarded, the very low rheumatic morbidity in tropical-rainy climate is noted, whereas it is very high in temperate-rainy zones, with a medium position in dry climate. (E) Rheumatism has decreased since 1947. The cause of this phenomenon can be attributed to improved living conditions in the Mexican Republic. (F) In Mexico rheumatic heart disease produces about one-third of the cases of cardiopathy. Therefore, a special campaign should be organized to prevent rheumatism and, in particular, the heart complications of this disease.

AUTHORS' SUMMARY.

FREYCHE, M. J.; PAYNE, A. M. M. AND LEDERREY, C.: POLIOMYELITIS IN 1953. (Bulletin World Health Organization, 12:595, 1955).

The world incidence of poliomyelitis has been reviewed on the basis of the available statistical data. In Africa, 1953 saw a rise in the apparent incidence of the disease in the Middle Congo (French Equatorial Africa), Morocco (French Zone), Ruanda-Urundi, Tanganyika, Tunisia, The Union of South Africa, Upper Volta (French West Africa), and, in particular, in Angola and in Egypt. The rise in the latter country is believed to have been due largely to improved reporting. The figures for Kenya and Southern Rhodesia were lower than those for 1952, and there was a still more marked improvement in the situation in Mauritius and Uganda. In America, Canada suffered the worst poliomyelitis epidemic in its history; Argentina, Rio de Janeiro and the State capitals of Brazil, Colombia, El Salvador, Greenland, Mexico, Nicaragua and Uruguay were all more or less seriously affected, but there was a substantial drop in apparent morbidity in Cuba, the U.S.A. and, probably, Cuba. The available data with regard to Asiatic countries are in most cases too incomplete to enable

any conclusions to be drawn. In Europe, Sweden suffered the most serious outbreak ever recorded there, and Finland was also severely affected. Both Austria and Switzerland suffered a relatively high incidence, and an upward trend was observed in France, Ireland, Norway, U.K. and 4 southern European countries. Elsewhere the incidence was generally lower than in previous years. The information available regarding the virus types responsible for clinical poliomyelitis in most parts of the world is so fragmentary that no definite conclusions are justifiable. The total number of polioviruses isolated and typed from clinical cases since the different types were recognized is probably not much more than 3,000-4,000. The results for about 2,200 strains are given in this study and of these 84 per cent are type 1, 7 per cent type 2, and 9 per cent type 3. This does not represent the relative prevalence of the different types. Serological studies have made it clear that all three types are widely distributed and that, as a generalization, antibodies are acquired to all three types at about the same rate. It is now widely recognized that many viruses other than poliovirus can cause the clinical picture of nonparalytic poliomyelitis, and that considerable proportions of such cases are not due to poliovirus infection. Evidence is also accumulating which suggests that there are viruses other than the three recognized types of poliovirus which can cause the clinical picture of paralytic poliomyelitis, and that they may be more common than has been thought.

MICHAEL A. BRESCIA, M.D.

RHOADS, P. S.; SIBLEY, J. R. AND BILLINGS, C. E.: BACTEREMIA FOLLOWING TONSILLECTOMY. EFFECT OF PREOPERATIVE TREATMENT WITH ANTIBIOTICS IN POSTOPERATIVE BACTEREMIA AND IN BACTERIAL CONTENT OF TONSILS. (Journal American Medical Association, 157:877, March 12, 1955).

Blood cultures taken just after tonsillectomy were positive in 28.3 per cent of a group of 68 patients who received no antibiotic therapy prior to tonsillectomy. The incidence of bacteremia was reduced to 5.9 per cent in a group of 20 subjects who received penicillin in a daily dose of 600,000 to 800,000 U.I.M. for 4 to 10 days prior to tonsillectomy. The incidence of bacteremia in a group of 29 patients who received 600,000 to 800,000 U. (half

this dose for children) of procaine penicillin 12 to 18 hours and 1 hour prior to tonsillectomy or in a small group (7) of patients receiving 900,000 to 1,200,000 U. orally daily for 5 to 7 days prior to operation was not reduced below that of the control group. B. hemolytic streptococci were obtained from blood cultures 4 times, pneumococci once, A. hemolytic streptococci (green forming) or gamma anhemolytic streptococci 28 times, and a combination of B. hemolytic streptococci and gamma anhemolytic streptococci twice. The need for several days preoperative treatment with penicillin to prevent posttonsillectomy bacteremia is obvious. In the control series of 68 patients who had no antibiotic treatment immediately preceding tonsillectomy, B. hemolytic streptococci were present in 57.4 per cent of the cultures of the excised tonsils, although these micro-organisms were found in only 28.26 per cent of throat cultures taken just before the operation. Among patients receiving penicillin in single doses only the day before and the day of tonsillectomy, 31.03 per cent had B. hemolytic streptococci in the excised tonsils, although these micro-organisms were not present in the throat cultures taken just before tonsillectomy. B. hemolytic streptococci were found only once in cultures from the excised tonsils of persons receiving penicillin IM each day for 4 to 10 days prior to tonsillectomy. Most of the gram-positive micro-organisms except micrococci and Gaffkya tetragenus, were greatly reduced in number by penicillin administered IM, but gram-positive micro-organisms, such as K. pneumoniae, Aerobacter aerogenes and E. coli, were found in increased numbers in cultures from the throats and excised tonsils of these subjects.

AUTHORS' SUMMARY.

GIERSON, H. W. AND MARX, J. I.: TUBERCULOUS MENINGITIS. THE DIAGNOSTIC AND PROGNOSTIC SIGNIFICANCE OF SPINAL FLUID SUGAR AND CHLORIDE. (Annals of Internal Medicine, 42:902, April 1955).

A series of 231 cases of tuberculous meningitis observed at the Los Angeles County General Hospital has been analyzed to ascertain the diagnostic and prognostic significance of the spinal fluid sugar and chloride levels. Both factors were depressed in most instances, but in a significant number of cases one or the other

was normal at the time of the initial examination and remained so throughout the diagnostic period. In our series the sugar levels failed to reflect the meningeal tuberculosis more often than did that of the chloride. Neither the spinal fluid glucose nor chloride provides a sensitive prognostic index. But again, of the two we find the chloride to be somewhat more reliable. It is therefore recommended that chloride determinations be part of the routine spinal fluid analysis in tuberculous meningitis. AUTHORS' SUMMARY.

ACUTE POLYARTHRITIS AND INFECTIONS OF THE UPPER RESPIRATORY TRACT. PROPHYLAXIS (Semaine hôp. Paris, 31:490-500, Feb. 6, 1955). This study is a review of the literature combined with personal observations. In rheumatic fever, prevention of rheumatismal relapses is the indispensable complement of hormone treatment of the acute attack. Prevention may be accomplished in three ways: 1. Immediate treatment with antibiotics, especially penicillin and secondarily chlortetracycline, of angina and streptococcic respiratory infections. This treatment in patients with long-standing rheumatism yields a decrease in relapses by comparison with nontreated patients, but penicillin therapy is inadequate in these patients with disease of long duration. 2. For continuous prophylaxis penicillin is preferable to the sulfonamides; continuous antibiotic administration is begun at the same time as hormone therapy, that is, immediately. Penicillin is injected intramuscularly for the first few days and is then given orally during the next years. This daily treatment decreased the number of rheumatic relapses in the proportion of one to five at least in two hospitals. Injectable preparations of penicillin with delayed action seem to be promising. 3. The suppression of residual foci of infection, such as in the tonsils or teeth, is sometimes indispensable. Lastly, it is pointed out that acute infectious arthritis of rhinopharyngeal origin, aside from Bouillaud's disease, also benefits from antibiotic prophylactic therapy.—*J.A.M.A.*

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